

Atlanto Axial Rotatory Dislocation in Adults: A Rare Complication of an Epileptic Seizure—Case Report

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Abstract

Atlanto Axial Rotatory Dislocations (AARDs) are a heterogeneous group of post-traumatic pathologies typical of the pediatric age, and rare in adults. We describe the case of a 34-year-old woman, developing Atlanto Axial Rotatory Fixation (AARF) after a generalized tonic-clonic epileptic seizure, an extremely rare traumatic cause never described in literature. AARF was detected only 1 month after the accident and nonsurgical treatment was attempted at the beginning. The patient underwent surgery only 2 months after the accident. The best treatment should be conservative reduction within 1 month; when it is not possible, it is advisable to perform surgery as soon as possible. C1–C2 fixation with Harm's technique is the gold standard for fixed luxations. Delay of treatment makes intraoperative reduction more difficult and increase the establishment of the chronic permanent change of neck muscles and ligaments.

Key words: atlanto axial dislocations, Atlanto Axial Rotatory Dislocations, Atlanto Axial Rotatory Fixation, cervical luxation, tonic-clonic seizures

Introduction

Atlanto Axial Rotatory Dislocations (AARDs) are rare conditions resulting from extreme rotation and distraction trauma of the cervical spine. They occur more commonly in children from 0 to 18 years in whom there is a high degree of concordance for rotational behavior of C1 and C2,¹⁾ weakness and malleability of periarticular soft tissue and increased mobility of the articular joints. Occasionally they are seen in adults, especially when pre-existing C1–C2 instability is present (as in rheumatoid arthritis, Down syndrome, and an assortment of congenital cervical anomalies.¹⁾ The terminology associated with traumatic rotatory injuries at this level is somewhat confusing and a clear distinction should be made. AARDs are a heterogeneous group of C1–C2 rotatory luxation also called Atlanto-Axial Rotatory Subluxation (AARS) if the luxation is reducible with a corrective counter-rotation of the neck, and Atlanto-Axial Rotatory Fixation (AARF) if the luxation is fixed. AARS can be treated usually with a conservative treatment^{2,3)} but in AARF surgery may be necessary. We describe the case of a 34-year-old woman developing an AARD after a generalized tonic-clonic epileptic seizure, an extremely rare traumatic cause of AARD, never described in literature.

Case Report

A 34-year-old woman was referred with a history of drug-resistant epilepsy since adolescence, and was treated with temporal lobectomy and poly-drug therapy. After a tonic-clonic seizure the patient presented with neck pain with the head in “cock robin” position (chin turned toward right side and the neck laterally flexed to the left side). She was treated at the beginning as a torticollis with nonsteroidal anti-inflammatory drugs (NSAIDs) and Schanz collar. One month later, radiological exams were performed and AARD was detected. Computed tomography (CT) scan (Fig. 1) documented a left rotation of the atlas posterior arch and left atlanto-axial facet locking without anterior atlanto-dental gliding. Magnetic resonance imaging (MRI) scan showed integrity of transverse ligament, hyperintensity of the alar ligaments in short time inversion recovery (STIR) and T₂ sequences and a normal course of the vertebral arteries on magnetic resonance (MR) angiography (Fig. 2). According to Fielding and Hawkins this was “type 1” luxation.⁴⁾ C1–C2 rotation angle was about 30 degree. We attempted to achieve reduction with nonsurgical treatment. The patient was treated with cervical tractions, Philadelphia neck brace, and 8 mg/die thiocolchicoside. As conservative treatment failed to reduce the deformity, the patient underwent surgery after 1 month. Posterior approach was preferred. After muscles dissection and

C1–C2 atlanto-axial membrane excision, the left atlanto-axial joint was detected and the articular cartilage on the joint surfaces was denuded with a microdrill. C1 lateral mass screws and C2 isthmus screws were positioned with C1–C2 Goel-Harms technique. Then, with a combination of distraction, derotation, and extension,

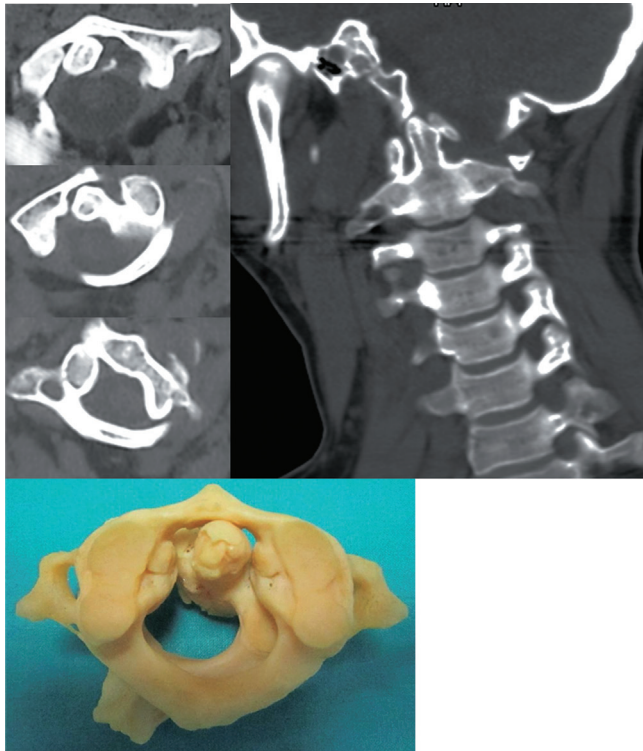


Fig. 1 Computed tomography scan and model reconstruction show the Atlanto Axial Rotatory Fixation.

the joint surfaces were completely relocated obtaining a good intraoperative reduction. Finally, fixation with rods was performed (Figs. 3, 4). Pain remitted after surgery and the neck returned to the physiological position. The patient was on Philadelphia brace for 3 months and Schanz collar for 3 months further. X-ray scan after 3 months and 6 months from surgery revealed good realignment of the luxation. After 1 year, she slightly tended to incline the neck on the right side. She had no pain and the stabilization system was correctly located (Fig. 5).

Discussion

AARDs are post-traumatic pathologies resulting from rotatory traumas holding on C1–C2 bone-ligament complexes. Physiologically, the C1–C2 joint is responsible for as much as 60% of the total rotation of the neck behaving as the main rotational pivot of the cervical spine. There are 6 degrees of freedom on the atlanto-axial joint: (1) angular motion (flexion and extension); (2) rotation (right and left); and (3) linear motion and translation (anterior and posterior, right and left). Although the upper cervical spine is the most mobile segment of the whole spinal column, the stability of C1–C2 joint is ensured by the atlas transverse ligament having high resistance against angular motion (flexion and extension), by the alar ligaments limiting lateral flexion movements and by the joint capsules limiting rotatory movements. The resistance against rotation strength on the lower cervical spine is for most depending from the annular fibers of the intervertebral disc, but since there is no disc in C1–C2, rotation stress only bear down on the atlanto-axial joints and C0–C1 joints. When they are overstressed by rotatory movements, there

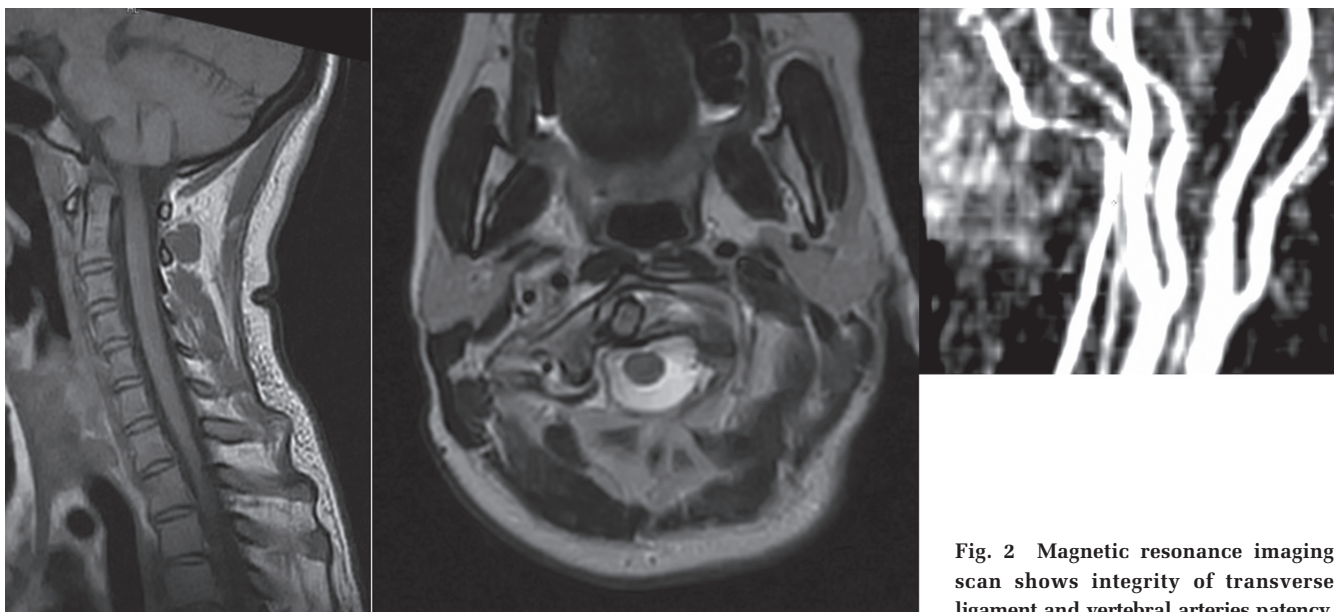


Fig. 2 Magnetic resonance imaging scan shows integrity of transverse ligament and vertebral arteries patency.

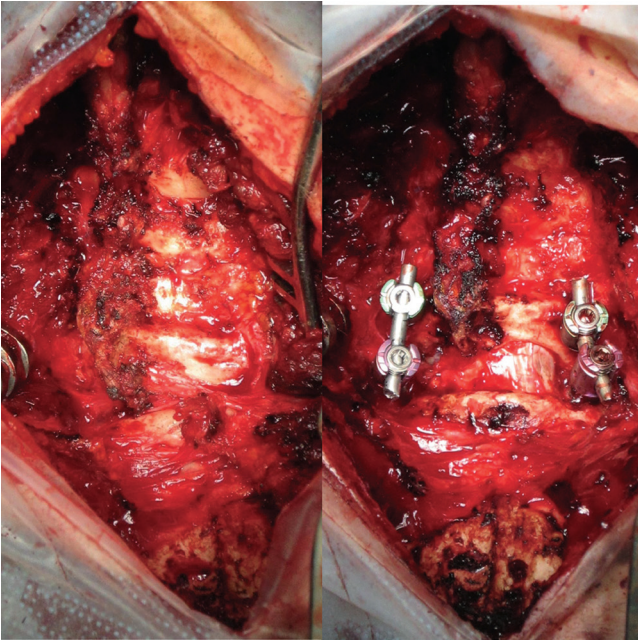


Fig. 3 Harms fixation and realignment of the luxation.

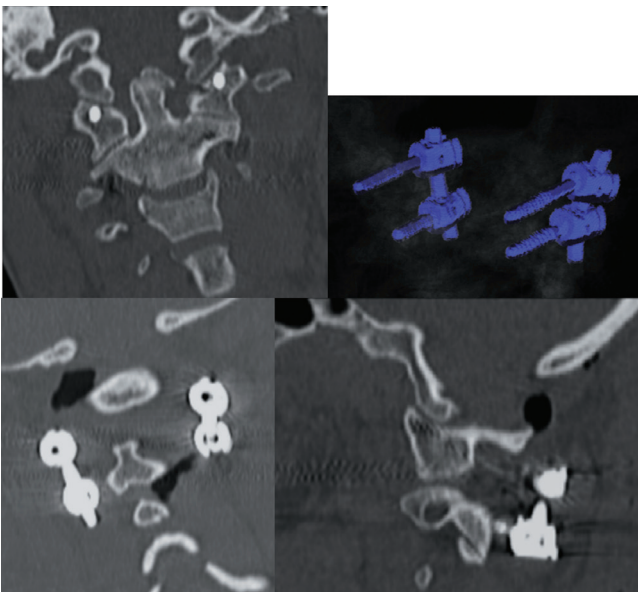


Fig. 4 Postoperative computed tomography scan.

are more chances of developing instability and capsule distraction, and consequently AARS. To have a luxation the abnormal critical separation angle between C1–C2 was defined by 18 degrees and 20 degrees.¹⁾ Young children have more elastic and slack joints ligaments, and also weak periarticular soft tissue. This may cause a worse lateral tilt during an extreme forward rotation but at the same time it may explain the success of conservative treatments as bed rest, muscle relaxant drugs, and eventually neck braces.²⁾ In adult patients, to have a C1–C2 luxation high kinetic

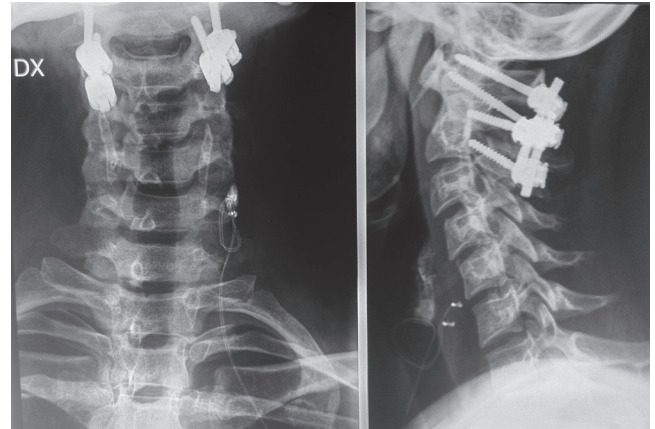


Fig. 5 X-Ray (XR) scan 1 year after surgery.

energy is needed to exceed the elastic forces of the joints. A violent rotatory trauma may cause injuries and tears to the capsule, with the consequence of intense pain and muscular spasm; if not promptly treated this condition may lead to irreducible AARF. Aetiology is usually traumatic, and car accidents represent the main cause, followed by domestic incident and school traumas.²⁾ In a number of cases⁵⁾ it is associated with nasopharyngeal infections (Grisel's syndrome), though the correct pathogenesis is still unclear. Cases have been described in patients with otitis media, retropharyngeal abscesses, lupus, rheumatoid arthritis, Down syndrome, Morquio disease, and in patients who have undergone otolaryngologic procedures.^{1,6)} Some authors described AARDs associated with cervical dystonia.⁷⁾ Thus three factors probably contribute to the AARDs: (1) distraction and extreme rotation of the head, (2) loss of the protective function of the neck musculature, and (3) periarticular tissue laxity. Generalized tonic-clonic seizures cause repeated rotatory traumas on articular capsules without any muscular control weakening the joints and finally resulting in AARS. The delay in reduction and the cervical muscle spasm caused permanent changes that set the fixation. It would explain the failure of conservative treatment. Delayed or missed diagnosis can occur especially in adults and lead to severe consequences.⁸⁾ In a patient with persistent torticollis (particularly if is post-traumatic) if there is spasm of contralateral sternocleidomastoid muscle a high index of suspicion of AARD must be posed. CT scan must be performed to diagnose and classify the dislocation⁹⁾ and evaluate C0–C1 and C1–C2 joints. MRI scan is needed to evaluate transverse ligament, the articular synovial fluid, and periarticular edematous phenomena. MR angiography is needed to evaluate the exact status of vertebral arteries. Many surgical approaches are described for the treatment of AARDs.^{10–13)} The posterior approach with the Harms fixation¹⁴⁾ is the gold standard in fixed luxations (AARF) achieving a complete realignment and a correct stabilization. It is useful

to allow an intraoperative reduction distracting directly on the screw heads. Moreover in order to perform transarticular fixations (posterior Magerl's or anterior Dickman's technique)^{11,15,16)} either C1 and C2 facets have to be already aligned then they could be used only after reduction. Harms fixation system should be removed while we decided not to in order to prevent injuries following a new traumatic seizure. Delay in reduction more than 1 month in adult result in more difficult intraoperative reduction and could have clinical consequences. Our patient underwent surgery 2 months after the accident. She presented 1 year after reduction with light neck inclination to the right side, although she was symptom-free, the rotatory axis was maintained and the stabilization system was correctly located. Probably it was related to the opposite side sternocleidomastoid spasm, to the nuchal muscles spasm and to the chronic adhesive fibrotic changes in the articular surfaces, may be even worsened because of the delay in reduction. Delayed reduction more than 1 year can cause chronic insufficiency of transverse atlantal ligament with increase of atlanto-dens interval.¹⁾

Conclusion

AARDs in adults are rare, and can occur even after a violent generalized tonic-clonic epileptic seizure. The best treatment should be conservative reduction within 1 month; when it is not possible, it is advisable to perform surgery as soon as possible, in order to reduce the dislocation and stabilize the joint. C1–C2 fixation with Harm's technique is the gold standard for reduction and stabilization allowing to perform distraction maneuvers directly on screw heads. Delay of treatment makes intraoperative reduction more difficult and increases the establishment of the chronic permanent change of neck muscles and ligaments.

Conflicts of Interest Disclosure

The authors declare that they have no conflicts of interest, ethical adherence, and any financial disclosures. Pasquale Donnarumma and Alessandro Landi analyzed and interpreted the patient data regarding the primary disease and the follow-up. Pasquale Donnarumma and Paolo Missori are the major contributors in writing the manuscript. Roberto Tarantino, Nicola Marotta, and Roberto Delfini performed the surgery. All authors read and approved the final manuscript.

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